JACC Vol. 6, No. 1
July 1985:49–54

Surgical Repair of Uncomplicated Atrial Septal Defect Without "Routine" Preoperative Cardiac Catheterization

CLARENCE SHUB, MD, FACC, A. JAMIL TAJIK, MD, FACC, JAMES B. SEWARD, MD, FACC, DONALD J. HAGLER, MD, FACC, GORDON K. DANIELSON, MD, FACC

Rochester, Minnesota

Whether all patients with atrial septal defect should undergo cardiac catheterization before surgical correction is controversial. Of 152 patients of all ages with surgically documented isolated atrial septal defect (ostium secundum, ostium primum and sinus venosus types) who underwent preoperative two-dimensional echocardiography between January 1978 and December 1983, there were 55 (36%) (group 1) who did not have preoperative cardiac catheterization. These 55 patients are compared with the 97 patients (64%) who did have preoperative catheterization studies (group 2). Group 1 patients were younger (mean age 22 versus 36 years) and did not have clinical evidence of other associated cardiac disorders. Forty-four (80%) of the group 1 patients had typical findings for atrial septal defect on cardiac examination, chest radiograph and electrocardiogram compared with 16 (16%) of group 2 patients ($p < 0.001$). The most frequent reasons for cardiac catheterization in group 2 patients were documentation of diagnosis, usually because of atypical clinical findings, and exclusion of associated cardiac abnormalities. Contrast echocardiography, radionuclide shunt studies and Doppler echocardiography were used as additional confirmatory tests in 36 patients (65%). In 19 patients (35%), two-dimensional echocardiography was the only confirmatory test. There were no false positive two-dimensional echocardiographic studies.

There were no operative deaths or significant perioperative complications in any of the patients. At a mean follow-up of 28 months (range 6 to 64), there was one late death (3½ years postoperatively) in a 61 year old man with chronic congestive heart failure. It is proposed that, in carefully selected patients of all ages with typical clinical findings of atrial septal defect and confirmation of the diagnosis by two-dimensional echocardiography, surgical repair can proceed without preoperative cardiac catheterization.

(J Am Coll Cardiol 1985;6:49–54)

Atrial septal defect is a common congenital abnormality that can usually be diagnosed clinically. Patients with atrial septal defect often undergo preoperative cardiac catheterization primarily to confirm the diagnosis and to exclude associated cardiac abnormalities. However, if routine preoperative cardiac catheterization could be avoided with safety, at least in selected patients, this would have potentially important implications, especially in terms of patient comfort and cost. Such an approach has been used at the Mayo Clinic for more than 20 years (1) and has been recommended for children (2,3), but it has not gained widespread acceptance. Most cardiologists and surgeons still are reluctant to recommend operation without confirmatory data from a cardiac catheterization study.

Two-dimensional echocardiography, especially when supplemented by contrast or Doppler echocardiography, has proved to be reliable in the diagnosis of atrial septal defect and the assessment of associated hemodynamic and structural abnormalities of the heart (4). In selected patients at our institution, two-dimensional echocardiography has essentially supplanted cardiac catheterization as the confirmatory test for atrial septal defect. In a recent preliminary report (5), we described the clinical characteristics, operative outcome and role of two-dimensional echocardiography in the preoperative evaluation of 39 patients of various ages who underwent successful operative repair of atrial septal defect without preoperative cardiac catheterization. We now report our updated experience in a larger group of patients and compare this group with patients who did undergo cardiac catheterization.

From the Division of Cardiovascular Diseases and Internal Medicine, Division of Pediatric Cardiology and Section of Thoracic and Cardiovascular Surgery, Mayo Clinic and Mayo Foundation, Rochester, Minnesota. Manuscript received November 13, 1984; revised manuscript received February 12, 1985, accepted March 1, 1985.

Address for reprints: Clarence Shub, MD, Mayo Clinic, 200 First Street SW, Rochester, Minnesota 55905.

0735-1097/85/$3.30

JACC Vol. 6, No. 1
July 1985:49–54

Methods

Study patients. This retrospective analysis included all patients (n = 208) with surgically documented, isolated atrial septal defect who underwent preoperative two-dimensional echocardiography between January 1978 and December 1983. The technical details and success rate of visualizing atrial septal defects utilizing the subcostal format have been previously described (4). Fifty-six of the 208 patients underwent cardiac catheterization at other institutions before referral to the Mayo Clinic and therefore were excluded from further analysis. Of the remaining 152 patients, 55 (36%) underwent surgical repair of atrial septal defect without preoperative cardiac catheterization (group 1). The 97 patients who did undergo preoperative cardiac catheterization at the Mayo Clinic constitute group 2.

Clinical diagnostic criteria for atrial septal defect. The clinical findings, electrocardiogram and chest radiograph were classified as being either typical or atypical for atrial septal defect on the basis of generally accepted criteria (6–8). For the purposes of this analysis, typical physical findings included a wide, persistently split second heart sound and a systolic ejection murmur heard along the left sternal border. A right ventricular conduction delay on the electrocardiogram with either right-axis deviation (clockwise, inferiorly directed QRS loop) or vertical QRS frontal plane axis was considered typical for ostium secundum atrial septal defect. No frontal plane QRS axis criteria were required for sinus venosus defect (9). Left-axis deviation (counterclockwise, superiorly directed QRS loop) was considered typical for ostium primum defect. Typical radiographic criteria included a prominent main pulmonary artery segment, increased pulmonary vasculature and mild cardiomegaly.

Follow-up. Anatomic findings at operation, perioperative complications and postoperative follow-up were obtained by review of surgical and clinical records and by letter or telephone call to each patient. Follow-up was completed in 51 group 1 patients (93%); mean follow-up time was 28 months (range 6 to 64). Four patients were lost to follow-up. Differences between groups were analyzed by chi-square analysis.

Results

Group 1 (no preoperative catheterization)

Patients. The 55 patients in group 1 included 20 men and boys (36%) and 35 women and girls (64%), for a female to male ratio of 1.8:1 (Table 1). The mean age was 22 years (range 1 to 67). Forty-three patients (78%) were less than 40 years of age and 29 (53%) were less than 20 years of age; only 4 were greater than 50 years of age. No patient had a history of angina pectoris or prior myocardial infarction. Forty patients (73%) had an ostium secundum defect, 8 (14%) had an ostium primum defect and 7 (13%) had a sinus venosus defect.

Table 1. Characteristics of 152 Patients Studied

	Group 1	Group 2	p Value*
Number of patients	55	97	—
Age (yr)			
Mean	22	36	< 0.001
Range	1 to 67	0.25 to 79	
Female/male ratio	1.8:1	1.2:1	NS
Type of atrial septal defect			
Secundum	40 (73%)	73 (75%)	NS
Primum	8 (14%)	11 (11%)	NS
Sinus venosus	7 (13%)	13 (14%)	NS
Typical clinical findings present			
Cardiac examination	49 (89%)	42 (43%)	< 0.01
Electrocardiogram	52 (95%)	53 (55%)	< 0.001
Chest radiograph	53 (96%)	52 (54%)	< 0.001
All three	44 (80%)	16 (16%)	< 0.001

*Analyses by two-sample *t* test.

Clinical characteristics. All three clinical criteria (cardiac examination results, electrocardiogram and chest radiograph) were considered to be typical for atrial septal defect in 44 patients (80%), and two of the three criteria were met in the remaining 11 patients. One or more additional noninvasive confirmatory tests were performed on 7 of these 11 patients (Table 2). Typical cardiac findings were noted in 49 patients (89%), the electrocardiogram was typical in 52 (95%) and the chest radiograph was typical in 53 (96%). Atypical findings are listed in Table 2.

Echocardiography. Although the actual defect was not visualized in all patients (Table 3), each patient had the echocardiographic pattern of right ventricular volume overload and right atrial enlargement, features thought to be consistent with the diagnosis of atrial septal defect (4). Forty additional positive confirmatory tests (radionuclide shunt study, two-dimensional contrast study and Doppler echocardiography) (Table 3) were obtained in 36 patients (65%); in 19 patients (35%) the two-dimensional echocardiogram was the only confirmatory test. Doppler studies performed at the time of echocardiography were the most frequently obtained tests. These were obtained primarily in 1982 and 1983 and were used most often in younger patients. All contrast echocardiographic studies were performed before 1983 (4).

There were no false positive cases; that is, no patient came to operation without a confirmed atrial septal defect. When seen, the defect was correctly classified as ostium secundum, ostium primum or sinus venosus in 40 of 43 patients (93%). In three patients, echocardiographic misclassification occurred: 1) an 11 year old girl studied in 1981 had a very large (5 cm) ostium secundum defect at operation (extended superior to the fossa ovalis) but was thought to have a "combined" sinus venosus and secundum defect by

Table 2. Confirmation of Diagnosis in 11 Patients in Group 1 With Atypical Findings

Year	Age (yr) & Sex	Type of Defect	Atypical Findings	Echocardiogram		Radionuclide Shunt Study	Comment
				Contrast	Doppler		
1978	31M	Secundum	CXR normal	–	–	+	Qp:Qs = 1.8
1980	1.3M	Secundum	S_2 normal	–	–	–	
1980	5F	Secundum	? PS on examination	–	–	–	PV normal on echocardiogram and at operation
1980	29F	Secundum	ECG normal	–	–	+	Qp:Qs = 3.0
1980	20F	SV	RAD only	+	–	–	
1981	39M	SV	CXR normal	+	–	+	Qp:Qs = 1.8
1981	4F	Secundum	MR on examination	–	–	–	Cleft MV on 2D echocardiogram; confirmed at operation
1982	19F	Secundum	RAD only	–	+	–	
1983	43M	Secundum	S_2 normal	–	+	–	
1983	3M	Secundum	S_2 normal	–	–	–	
1983	46M	SV	S_2 indeterminate	–	+	–	

CXR = chest radiograph; ECG = electrocardiogram; F = female; M = male; MR = mitral regurgitation; MV = mitral valve; PS = pulmonary stenosis; PV = pulmonary valve; Qp:Qs = pulmonary to systemic flow ratio; RAD = right-axis deviation; S_2 = second heart sound; SV = sinus venosus; 2D = two-dimensional; + = study performed; – = study not performed.

echocardiography; 2) a 14 year old boy studied in 1978 had a sinus venosus defect at operation that was not visualized by echocardiography, but he also had a small ostium secundum septal defect; and 3) a 3 year old girl studied in 1982 had an ostium secundum defect, but the echocardiographer could not distinguish between a posterior ostium secundum defect and a sinus venosus defect. None of these misclassifications of atrial septal defect affected the surgical outcome.

Seven patients had a partial anomalous pulmonary venous connection (six patients with a sinus venosus atrial septal defect and one 23 year old woman with an ostium secundum defect) that was not diagnosed preoperatively by two-dimensional echocardiography. In each case, the anomalous connection was found at operation and repaired. One patient, a 6 year old girl with an ostium secundum defect, was found to have minor pulmonary stenosis that was not appreciated on echocardiography but was identified at operation; valvotomy was performed.

Perioperative complications and follow-up. There were no operative deaths or significant perioperative complications in any of the group 1 patients. In the follow-up period after hospital discharge (mean follow-up 28 months), one death occurred in a 61 year old man who had had successful repair of a sinus venosus defect but had a history of preoperative congestive heart failure and chronic atrial fibrillation. This patient was receiving digitalis, furosemide and quinidine. Although his condition was improved postoperatively, he died suddenly 3½ years postoperatively. A 1 year old girl with ostium primum defect who also had suture

Table 3. Visualization of Atrial Septal Defect and Additional Positive Confirmatory Tests in Group 1 According to Age

	Patient Age Groups (yr)							
	0 to 9	10 to 19	20 to 29	30 to 39	40 to 49	50 to 59	60 to 69	Total
Total no. in age group	23	7	7	4	10	2	2	55
Defect visualized	23	5	5	2	6	1	0	42
Radionuclide shunt study	0	1	1	2	5	0	2	11
Doppler echocardiography	8	3	1	1	3	0	0	16
Contrast echocardiography	1	2	7	1	1	0	1	13

JACC Vol. 6, No. 1
July 1985:49–54

repair of a cleft mitral valve underwent mitral valve replacement 6 months later for recurrent severe mitral regurgitation. Mild residual asymptomatic mitral regurgitation was noted in three other patients who had undergone repair of ostium primum atrial septal defect. All other patients were in functional class I (New York Heart Association criteria), and no additional cardiac diagnoses were made.

Group 2 (patients with preoperative cardiac catheterization)

Clinical characteristics (Table 1). Group 2 patients were significantly older than patients in group 1. There were 44 patients (45%) older than 50 years, 18 (19%) older than 60 years and 3 older than 70 years. Thirty-one patients (32%) were less than 20 years old. The ratio of female to male patients was lower than in group 1, but this difference was not statistically significant. Inconsistencies among the three clinical variables were more common. Findings typical for atrial septal defect were much less frequent in group 2 than in group 1. Typical cardiac findings were found in only 43% of patients, typical electrocardiographic findings were present in 55% and the chest radiograph was typical in 53%; all three criteria were met in only 16%.

Indications for cardiac catheterization (Table 4). Cardiac catheterization was performed most frequently in this group (53%) to document the diagnosis of atrial septal defect. In 46 (90%) of these 51 patients the clinical assessment was inconsistent and one or more atypical clinical findings for atrial septal defect were present. In 38 patients (39%), cardiac catheterization was performed to exclude associated conditions, most frequently pulmonary hypertension or coronary artery disease. Thirteen patients underwent coronary angiography (8 men and 5 women) for suspected coronary artery disease; 12 had a history of angina. The mean age of this group was 54 years (range 29 to 70). Four patients had significant coronary artery disease; three of these underwent coronary artery bypass graft operation at the time of atrial septal defect repair. Other reasons for cardiac catheterization included assessment of valvular regurgitation (five patients) and shunt quantitation (three patients). Reasons for cardiac catheterization in the 16 patients with typical findings for atrial septal defect are shown in Table 5. Five patients underwent catheterization only for confirmation of diagnosis; in the remainder, specific clinical questions prompted the study.

There were no significant perioperative complications in group 2.

Table 4. Reasons for Cardiac Catheterization in Group 2

Reason	No.	Percent*
Document diagnosis of atrial septal defect	51	53
Atypical or inconsistent findings†	46	
Typical findings‡	5	
Exclude associated abnormalities	38	39
Coronary artery disease§	13	
Pulmonary hypertension	13	
Pulmonary stenosis	6	
Partial anomalous pulmonary venous connection	6	
Assess degree of associated valvular regurgitation	5	5
Mitral valve	4	
Ostium primum defect	2	
Ostium secundum defect	2	
Aortic valve	1	
Quantitate interatrial shunt	3	4

*Based on total of 97 patients in group. †Atypical findings in regard to one or more of the three criteria. ‡Typical findings in regard to all three criteria. §Three patients underwent concomitant coronary artery bypass graft operation.

Diagnostic Role of Two-Dimensional Echocardiography (groups 1 and 2)

In addition to confirming the diagnosis, two-dimensional echocardiography played a significant role in the diagnostic process for both groups of patients. Clinically unsuspected atrial septal defect was first diagnosed at the time of two-dimensional echocardiography and before cardiac catheterization in 17 patients (17%); 7 of these had a normal clinical examination, with normal findings or very subtle abnormalities on the electrocardiogram and chest radiograph. Before catheterization, the clinical diagnosis was altered in 13 patients on the basis of the two-dimensional echocardiographic findings. In 11 patients, the diagnosis of the specific type of atrial septal defect was changed; in 1 a diagnosis of total anomalous pulmonary venous connection was altered to ostium primum atrial septal defect and in 1 the clinical diagnosis of ventricular septal defect was changed to atrial septal defect.

Additional cardiac diagnoses that were unsuspected clinically and were first made at the time of echocardiographic examination (before catheterization), as well as additional diagnoses correctly excluded by echocardiography, are shown in Table 6.

Discussion

Repair of Atrial Septal Defect Without Preoperative Cardiac Catheterization

Clinical findings. This study documents that carefully selected patients with typical clinical findings of atrial septal defect in whom the diagnosis is confirmed by two-dimensional echocardiography can undergo operative repair safely without preoperative cardiac catheterization. The number of patients involved in this study (55) represents a substantial

Table 5. Reasons for Cardiac Catheterization in Patients With Typical Findings for Atrial Septal Defect

Age (yr)	Type of Defect	Reason	Comment
10 mo	Sec	Confirm diagnosis	Confirmed
1	Sec	? PS	No PS at catheterization
2	Sec	? PAPVC	PAPVC not found
6	SV	Exclude MR	No MR at catheterization
11	SV	? persistent LSVC, PAPVC	Persistent LSVC confirmed, no PAPVC
15	Prim	Confirm diagnosis	Confirmed
16	Prim	Confirm diagnosis	Confirmed
20	Sec	? PAPVC	PAPVC not found
29	Sec	Confirm diagnosis	Confirmed
39	Sec	? PH, ? MR	Echocardiographic diagnosis of MVP, mild MR at catheterization, no PH
45	Sec	? PS	No PS at catheterization
46	Sec	? PH	? PH on echocardiogram, no PH at catheterization
52	Sec	Assess coronary arteries	Normal coronary arteries at catheterization
53	Sec	Confirm diagnosis	Confirmed
56	Sec	Quantitate shunt	Qp:Qs = 3.2:1
66	SV	? PH	? PH on echocardiogram, no PH at catheterization

LSVC = left superior vena cava; MR = mitral regurgitation; MVP = mitral valve prolapse; PAPVC = partial anomalous pulmonary venous connection; PH = pulmonary hypertension; Prim = ostium primum; PS = pulmonary stenosis; Qp:Qs = pulmonary to systemic flow ratio; Sec = ostium secundum; SV = sinus venosus.

percent (36%) of the overall group of patients who underwent surgical repair of atrial septal defect at the Mayo Clinic from 1978 to 1983. Most of the patients were young adults with ostium secundum defect. However, children, older patients and patients with an ostium primum or sinus venosus defect were also included in this group. Approximately half (47%) of all patients less than 15 years of age did not undergo cardiac catheterization. This series is not directly comparable with the pediatric patient group recently reported by Freed et al. (3) (in which 69% of patients did not undergo catheterization). Patients with ostium primum defect were excluded from their series.

Echocardiographic findings. The echocardiographic diagnosis is best made by using the subcostal format in which the characteristics of the atrial septal defect can be best appreciated (4). Other indirect echocardiographic criteria supporting the diagnosis of interatrial shunt include right atrial and ventricular enlargement, abnormal septal motion and increased excursion of the tricuspid valve. In our series, none of the patients sent to operation without preoperative cardiac catheterization had a false positive diagnosis of atrial septal defect. Abnormalities that were missed by two-dimensional echocardiography—most commonly partial anomalous pulmonary venous connection, even though suspected in cases of sinus venosus atrial septal defect (4)—were readily identified at operation and successfully repaired in each instance.

Confirmation of diagnosis is possible in most patients (4). Ideally, the defect should be visualized directly. If not,

Table 6. Additional Cardiac Diagnoses by Two-Dimensional Echocardiography

Diagnosis	No.
Cardiac diagnoses not suspected clinically	
Mitral valve prolapse	6
Tricuspid valve prolapse	2
Atrial septal aneurysm	2
Hypertrophic cardiomyopathy	1
Tricuspid valve vegetation	1
Mitral stenosis	1
Tricuspid insufficiency	1
Aortic insufficiency*	1
Pulmonary insufficiency*	1
Left superior vena cava	1
Cardiac diagnoses excluded	
Ventricular septal defect	5
Pulmonary stenosis	3
Ebstein's anomaly	3
Dilated cardiomyopathy	1
Aortic stenosis	1
Cleft mitral valve	1

*By pulsed wave Doppler echocardiography.

JACC Vol. 6, No. 1
July 1985:49–54

an interatrial shunt should be clearly documented by Doppler echocardiography or contrast echocardiography before surgical repair. If any doubt exists, for example, because of inadequate images, equivocal Doppler signal or suspected partial anomalous pulmonary venous connection with an intact atrial septum, cardiac catheterization is indicated. At our institution, pulsed wave Doppler echocardiography has supplanted contrast echocardiography for additional confirmation of a left to right shunt. A radionuclide shunt study, to assess the magnitude of the shunt, is also rarely performed because clinical examination, electrocardiogram, chest radiograph and two-dimensional Doppler echocardiogram usually provide sufficient data.

Diagnosis of associated cardiac abnormalities. In the patients in this study who did not undergo cardiac catheterization, two-dimensional echocardiography was able to diagnose or exclude associated cardiac abnormalities in nearly all. "Missed" diagnoses, such as partial anomalous pulmonary venous connection in seven cases and mild pulmonary stenosis in one, did not substantially affect the operation or outcome. Identification of a sinus venosus atrial septal defect, which is frequently associated with partial anomalous pulmonary venous connection, is not considered to be an indication for cardiac catheterization. Inspection of pulmonary venous connections by the surgeon at operation is required, however. If a persistent left superior vena cava is suspected and the surgeon wishes to define systemic venous anatomy preoperatively, additional testing can be done.

Indications for cardiac catheterization. As expected, the clinical examination, electrocardiogram and chest X-ray film were more often typical (80 versus 16%) for atrial septal defect in the patients (group 1) without preoperative cardiac catheterization. In group 2, the diagnosis was often unclear or complicating features were present, such as pulmonary hypertension or coronary artery disease. Cardiac catheterization was used to document the diagnosis in 53% and to exclude associated abnormalities in 39%. Assessment of valvular regurgitation, another reason for cardiac catheterization in a smaller number of patients, may be accomplished by improved Doppler echocardiographic techniques.

Currently at the Mayo Clinic the most common indications for cardiac catheterization in patients suspected of having atrial septal defect are: 1) inconsistencies in the clinical data (atypical presentations); 2) assessment of associated cardiac abnormalities (for example, coronary artery disease or significant pulmonary hypertension); and 3) inadequate echocardiographic study.

On the basis of our experience, we propose that preoperative cardiac catheterization can be avoided in carefully selected patients who have typical clinical findings for atrial septal defect and whose diagnosis is confirmed by two-dimensional echocardiography. Further confidence is achieved with a confirmatory pulsed wave Doppler echocardiographic study. This approach has important implications in terms of lowering cost (3), avoiding the small but definite risk associated with cardiac catheterization, and patient comfort. To successfully practice this approach in the manner outlined, it is mandatory that a team approach be employed with appropriate interaction between the clinical cardiologist, echocardiologist and surgeon, each of whom must be experienced. After all three have agreed on the diagnosis of an uncomplicated atrial septal defect, the decision can then be made to proceed directly to operation without cardiac catheterization. The cardiac surgeon should be prepared to inspect pulmonary venous connections at the time of operation.

References

1. Pestana C, Weidman WH, Swan HJC, McGoon DC. Accuracy of preoperative diagnosis in congenital heart disease. Am Heart J 1966;72:446–50.
2. Neal WA, Moller JH, Varco RL, Anderson RC. Operative repair of atrial septal defect without cardiac catheterization. J Pediatr 1975; 86:189–93.
3. Freed MD, Nadas AS, Norwood WI, Castaneda AR. Is routine preoperative cardiac catheterization necessary before repair of secundum and sinus venosus atrial septal defects? J Am Coll Cardiol 1984;4:333–6.
4. Shub C, Dimopoulos IN, Seward JB, et al. Sensitivity of two-dimensional echocardiography in the direct visualization of atrial septal defect utilizing the subcostal approach: experience with 154 patients. J Am Coll Cardiol 1983;2:127–35.
5. Shub C, Tajik AJ, Seward JB, Hagler DJ, Danielson GK. Atrial septal defect: surgery without routine preoperative cardiac catheterization? (abstr). J Am Coll Cardiol 1983;1:651.
6. Perloff JK. The Clinical Recognition of Congenital Heart Disease. Philadelphia: WB Saunders, 1970:227–46.
7. DuShane JW, Weidman WH, Brandenburg RO, Kirklin JW. Differentiation of interatrial communications by clinical methods: ostium secundum, ostium primum, common atrium, and total anomalous pulmonary venous connection. Circulation 1960;21:363–71.
8. Leatham A, Gray I. Auscultatory and phonocardiographic signs of atrial septal defect. Br Heart J 1956;18:193–208.
9. Davia JE, Cheitlin MD, Bedynek JL. Sinus venosus atrial septal defect: analysis of fifty cases. Am Heart J 1973;85:177–85.